Raynaud phenomenon

Raynaud phenomenon is a condition in which the body's normal response to cold or emotional stress is exaggerated, resulting in abnormal spasms (vasospasms) in small blood vessels called arterioles. The disorder mainly affects the fingers but can also involve the ears, nose, nipples, knees, or toes. The vasospasms reduce blood circulation, leading to discomfort and skin color changes.

Raynaud phenomenon is episodic, meaning that it comes and goes. A typical episode lasts about 15 minutes after the cold exposure or stressor has ended and involves mild discomfort such as numbness or a feeling of "pins and needles." The affected areas usually turn white or blue when exposed to cold or when emotional stress occurs, and then turn red when re-warmed or when the stress eases.

Raynaud phenomenon is categorized as primary when there is no underlying disorder that accounts for the exaggerated response of the blood vessels. It is called secondary when it is associated with another condition. Secondary Raynaud phenomenon is often associated with autoimmune disorders, which occur when the immune system malfunctions and attacks the body's own tissues and organs. Autoimmune disorders with which Raynaud phenomenon can be associated include systemic lupus erythematosus, scleroderma, rheumatoid arthritis, and Sjögren syndrome.

Primary Raynaud phenomenon is much more common and usually less severe than secondary Raynaud phenomenon. In severe cases of secondary Raynaud phenomenon, sores on the pads of the fingers or tissue death (necrosis) can occur. Primary Raynaud phenomenon often begins between the ages of 15 and 25, while secondary Raynaud phenomenon usually starts after age 30. Some people with Raynaud phenomenon alone later go on to develop another associated condition; regardless of which comes first, these cases are classified as secondary Raynaud phenomenon.

Frequency

Raynaud phenomenon is a common condition, occurring in 3 to 5 percent of adults worldwide. It is more common in females than in males.

Causes

The signs and symptoms of Raynaud phenomenon are related to excessive narrowing (constriction) of small blood vessels in response to cold or stress. Variations in genes involved in this process may contribute to an individual's risk of developing Raynaud phenomenon, but the connection between these gene variations and the abnormal blood vessel response that occurs in this disorder is unknown.

Normally, blood vessels constrict and widen (dilate) in response to temperature changes and stress, a process called the vasomotor response. The nervous system and muscle cells in the walls of blood vessels control this response. When exposed to cold temperatures, blood vessels near the surface constrict to help keep warm blood in the body's core, near the vital organs. Blood vessels also constrict during stress as part of the body's "fight-or-flight" response, conserving the oxygen and heat carried by the blood for the body's basic functions. When the body is too warm, the surface vessels dilate to allow more blood to flow near the skin where it is cooler. Raynaud phenomenon is characterized by exaggeration of these normal vasomotor responses.

There are many causes of secondary Raynaud phenomenon. These include autoimmune disease; partial or complete loss of function of the thyroid gland (hypothyroidism); cancers of the blood, bone marrow, or immune system; disease processes that cause obstruction of blood vessels; exposure to certain medicines or chemicals; cigarette smoking; injury or trauma; prolonged repetitive motions such as typing; or long-term use of vibrating tools.

Inheritance Pattern

Raynaud phenomenon sometimes runs in families, but the inheritance pattern is unknown. Studies suggest that about 30 percent of people with a first-degree relative (parent, sibling or child) who has primary Raynaud phenomenon also have the condition.

Other Names for This Condition

- Raynaud disease
- Raynaud's
- Raynaud's disease
- Raynaud's phenomenon
- Raynaud's syndrome

Diagnosis & Management

Formal Diagnostic Criteria

- Belch J, Carlizza A, Carpentier PH, Constans J, Khan F, Wautrecht JC, Visona A, Heiss C, Brodeman M, Pécsvárady Z, Roztocil K, Colgan MP, Vasic D, Gottsäter A, Amann-Vesti B, Chraim A, Poredos P, Olinic DM, Madaric J, Nikol S, Herrick AL, Sprynger M, Klein-Weigel P, Hafner F, Staub D, Zeman Z. ESVM guidelines - the diagnosis and management of Raynaud's phenomenon. Vasa. 2017 Oct;46(6): 413-423. doi: 10.1024/0301-1526/a000661. Epub 2017 Sep 12. Review. *Citation on PubMed:* https://www.ncbi.nlm.nih.gov/pubmed/28895508

Formal Treatment/Management Guidelines

- Belch J, Carlizza A, Carpentier PH, Constans J, Khan F, Wautrecht JC, Visona A, Heiss C, Brodeman M, Pécsvárady Z, Roztocil K, Colgan MP, Vasic D, Gottsäter A, Amann-Vesti B, Chraim A, Poredos P, Olinic DM, Madaric J, Nikol S, Herrick AL, Sprynger M, Klein-Weigel P, Hafner F, Staub D, Zeman Z. ESVM guidelines - the diagnosis and management of Raynaud's phenomenon. Vasa. 2017 Oct;46(6): 413-423. doi: 10.1024/0301-1526/a000661. Epub 2017 Sep 12. Review. *Citation on PubMed:* https://www.ncbi.nlm.nih.gov/pubmed/28895508

Genetic Testing Information

- What is genetic testing? /primer/testing/genetictesting
- Genetic Testing Registry: Raynaud's disease https://www.ncbi.nlm.nih.gov/gtr/conditions/C0034734/

Research Studies from ClinicalTrials.gov

- ClinicalTrials.gov https://clinicaltrials.gov/ct2/results?cond=%22Raynaud+disease%22+OR+%22Raynaud+phenomenon%22+OR+%22Raynaud%27s%22+OR+%22Raynaud%27s+disease%22+OR+%22Raynaud%27s+phenomenon%22+OR+%22Raynaud%27s+syndrome%22+OR+%22paroxysmal+digital+cyanosis%22

Additional Information & Resources

Health Information from MedlinePlus

- Health Topic: Raynaud's Disease https://medlineplus.gov/raynaudsdisease.html

Additional NIH Resources

- National Heart, Lung, and Blood Institute (NHLBI): Raynaud's https://www.nhlbi.nih.gov/health-topics/raynauds
- National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS): Raynaud's Phenomenon https://www.niams.nih.gov/health-topics/raynauds-phenomenon

Educational Resources

- Canadian Centre for Occupational Health and Safety: Raynaud's Phenomenon https://www.ccohs.ca/oshanswers/diseases/raynaud.html
- Hospital for Special Surgery: Raynaud's Phenomenon https://www.hss.edu/condition-list_raynauds-phenomenon.asp

- Johns Hopkins Medicine Health Library: Raynaud's Phenomenon
  https://www.hopkinsmedicine.org/health/conditions-and-diseases/raynauds-phenomenon
- MalaCards: raynaud disease
  https://www.malacards.org/card/raynaud_disease
- Merck Manual Consumer Version: Raynaud Syndrome
  https://www.merckmanuals.com/home/heart-and-blood-vessel-disorders/peripheral-arterial-disease/raynaud-syndrome
- National Health Service (United Kingdom): Raynaud's
  https://www.nhs.uk/conditions/raynauds/

Patient Support and Advocacy Resources

- Lupus Foundation of America: About Raynaud's Disease
  https://www.lupus.org/resources/about-raynauds-disease
- Raynaud's Association
  https://www.raynauds.org/
- Scleroderma and Raynaud's UK
  https://www.sruk.co.uk/raynauds/what-raynauds/
- Scleroderma Foundation: Raynaud Phenomenon
  http://www.scleroderma.org/site/DocServer/Raynaud.pdf?docID=322

Scientific Articles on PubMed

- PubMed
  https://www.ncbi.nlm.nih.gov/pubmed?term=%28Raynaud+disease%5BTIAB%5D%29+AND+english%5Bla%5D+AND+human%5Bmh%5D

Catalog of Genes and Diseases from OMIM

- RAYNAUD DISEASE
  http://omim.org/entry/179600

Medical Genetics Database from MedGen

- Raynaud phenomenon
  https://www.ncbi.nlm.nih.gov/medgen/20474

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